

PULMONARY HYPERTENSION ASSOCIATED WITH LEFT HEART DISEASE

Definition

Patients have elevated pulmonary venous pressure (as reflected by a pulmonary capillary wedge pressure ≥ 15 mm Hg), usually as a consequence of either mitral or aortic valve disease or LV dysfunction.¹ It is therefore a post-capillary pulmonary hypertension, but with progression of the heart failure the combined post- and pre-capillary phenotype develops (Table 1).² Pulmonary hypertension related to left heart disease (LHD) by far represents the most common form of PH, accounting for 65–80% of cases.³

Pathophysiology

Chronic elevation of the LA or diastolic LV filling pressure causes a backward transmission of the pressure to the pulmonary venous system that triggers vasoconstriction in the arterial pulmonary bed (Figure 1). For reasons that remain unclear, some patients do not progress to reactive pulmonary vasoconstriction despite the presence of chronic advanced heart failure.¹ Pulmonary vascular disease in left heart disease is associated with a distinct pathophysiologic signature marked by greater exercise-induced lung congestion, arterial hypoxaemia, RV–PA uncoupling, ventricular interdependence, and impairment in O₂ delivery, impairing aerobic capacity.⁴

Diagnosis

Exertional dyspnea is the most common symptom. Diagnostic tests may reveal the causative condition and differentiate pulmonary venous hypertension from pulmonary arterial hypertension (PAH).

ECG LV rather than RV hypertrophy may be present.

Chest X-ray Pulmonary vascular congestion, pleural effusions and on occasion pulmonary edema.

Echocardiography Valve disease and reduced LV function can be detected. Left atrial enlargement, concentric remodelling of the LV (relative wall thickness >0.45), LV hypertrophy, and the presence of echocardiographic indicators of elevated LV filling pressure suggest PH due to left heart disease rather than PAH (Table 1). However, in the absence of detectable significant valve disease, Doppler echocardiography alone may not exclude PAH since pulmonary hypertension itself produces diastolic filling abnormalities in the LV.⁵

Chest CT Ground glass opacities and a mosaic perfusion pattern consistent with chronic

pulmonary edema, that are not seen in PAH.

Cardiac catheterization An elevated wedge pressure at end-expiration establishes the diagnosis but may not be always accurately measured, and may be normal due to the use of diuretics. Exercise or inotropic challenges may increase cardiac output and expose increased wedge pressure. Short-acting pulmonary vasodilators may also be used in cases with low wedge pressure and normal cardiac output.

Prognosis

Persistent valvular heart disease-pulmonary hypertension is an ominous disease that is almost universally associated with elevated pulmonary artery wedge pressure. Pulmonary vascular resistance is a major determinant of mortality in this condition and is related to diabetes mellitus and the residual effective area of the corrected valve.⁶ An elevated transpulmonary gradient (mean PAP minus mean PWP) >12 mmHg is suggestive of progressive intrinsic changes in the pulmonary circulation over-riding the passive increase in PWP.¹ This progressed form is also called reactive PH and resembles precapillary forms.

Therapy

It is targeting the underlying condition (Table 2). No heart failure drugs are contraindicated because of PH. Although preliminary results with phosphodiesterase type-5 inhibitors have been promising,^{7,8} pulmonary vasodilators may be harmful, and sildenafil has been detrimental in patients with persistent PH after successful correction of valvular heart disease.⁹ In the DYNAMIC trial, riociguat improved haemodynamics in patients with pulmonary hypertension and heart failure with preserved LV ejection fraction, but did not change clinical symptoms within the study period.¹⁰ Pulmonary artery denervation is a new promising option.¹¹

References

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